SUPPLEMENTS

Supplement 1. Publications on IBD course and follow-up in patients with a paediatric onset PSC.

Study	Design	Patients	Endoscopy at diagnosis	Treatment	Cancer
Shiau H et al.	Case-Control Retrospective	Cases: 39 IBD-PSC Controls: 95 UC Follow-up not reported	IBD-PSC vs IBD: *IBD-PSC higher rate of pancolitis (p=0.0009)*	IBD-PSC vs IBD: *IBD-PSC less steroids (p= 0.03)* *IBD-PSC less infliximab (p= 0.0011)* *IBD-PSC less surgery (p= 0.02)*	Data not reported
Lascurain et al. (1)	Case-Control Population-based Retrospective	Cases: 37 IBD-PSC (PSC-AIH 32%) Controls: 148 IBD Median follow-up 5 years	IBD-PSC vs IBD: *Higher rate of pancolitis (p=0.051)* *Similar rate of rectal sparing*	*IBD-PSC higher need of surgery (p=n.s.)* *IBD-PSC higher rate of pouchitis (p=n.s.)*	*1 CRC in IBD*
Ordonez et al. (2)	Case-Control Retrospective	Cases: 28 CAI (18 PSC, 6 PSC-AIH, 4 AIH, 2 CD, 2 HES) Controls: 27 UC Follow-up not reported	CAI vs UC: *Higher rate of pancolitis* *Similar rate of rectal sparing* *Similar rate of backwash ileitis*	*In CAI < need of steroids* *In CAI < need of azathioprine*	Data not reported
Faubion et al. (3)	Series Retrospective	36 PSC-IBD Follow-up not reported	*Pancolitis 80%* *Mild-moderate 83%* *Rectal sparing 13%*	*14% colectomy+pouch* *80% pouchitis*	*3 dysplasia:* *- 17 year-old* *- 23 year-old* *- 32 year-old*

PSC: primary sclerosing cholangitis, IBD: inflammatory bowel disease, CRC: colon rectal cancer, RS: rectal sparing, CAI: colitis associated autoimmune liver disease, AIH: autoimmune hepatitis, CD: Crohn´s disease, HES: hypereosinophilc syndrome, BWI: backwash ileitis.

Supplement 2. Baseline characteristics of IBD-PSC cases with and without overlap with AIH.

	PSC cases	PSC-AIH cases
Number	18	10
Males	11 (61%)	8 (80%)
Median age and 25-75th percentiles at IBD diagnosis in years	11, 9-15	15, 12-17
Median age and 25-75th percentiles at last follow-up in years	25, 21-35	30, 29-33
Median follow-up duration and 25-75th percentiles in years	14, 9-20	17, 13-21

PSC: primary sclerosing cholangitis, AIH: autoimmune hepatitis.

p significant < 0.05. For comparisons Fisher' Exact Test and Mann-Whitney Test were used when appropriate.

Supplement 3. Treatment for IBD during and at the last follow-up in IBD-PSC cases and IBD controls.

	Overall			At last follow-up		
	IBD-PSC N=28	**IBD-controls N=84**	**p**	**IBD-PSC N=28**	**IBD-controls N=84**	**p**
Not available	0 (0%)	2 (2.4%)		1 (3.6%)	3 (4%)	
Glucocorticoids	18/28 (64%)*	69/82 (84%)*	0.03	4/27 (15%)	8/81 (10%)	0.2
Thiopurine	13/28 (46%)	26/82 (32%)	0.1	5/27 (18%)	26/81 (32%)	0.4
TNF-alpha inhibitors	4/28 (14%)	23/82 (28%)	0.2	3/27 (11%)	7/81 (9%)	0.7
5-ASA compounds	26/28 (93%)	76/82 (82%)	1.0	18/27 (67%)	56/81 (69%)	0.8

PSC: primary sclerosing cholangitis, IBD: inflammatory bowel disease, TNF: tumour necrosis factor, ASA: aminosalicylic acid.

Supplement 4. Survival curve showing need of panproctolectomy with J-pouch/ileostomy in IBD-PSC cases and IBD-controls

Panproctocolectomy with J-pouch/ileostomy

p= 0.6
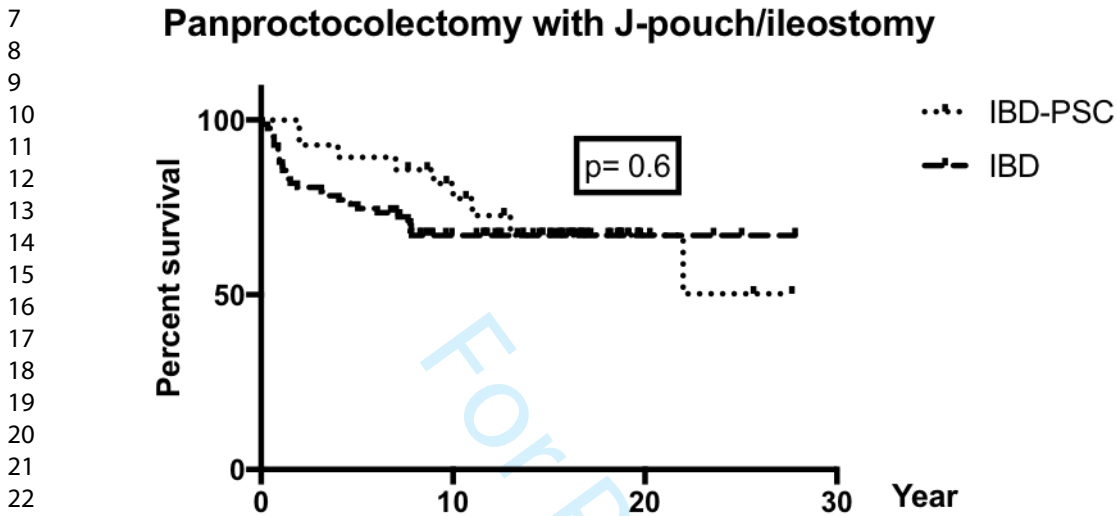
IBD-PSC
IBD

PSC: primary sclerosing cholangitis, IBD: inflammatory bowel disease

Supplement 5. Rate of panproctocolectomy with J-pouch reconstruction, pouchitis and recurrent pouchitis during the follow-up in IBD-PSC cases and IBD-controls

J-pouch surgery, pouchitis and recurrent pouchitis
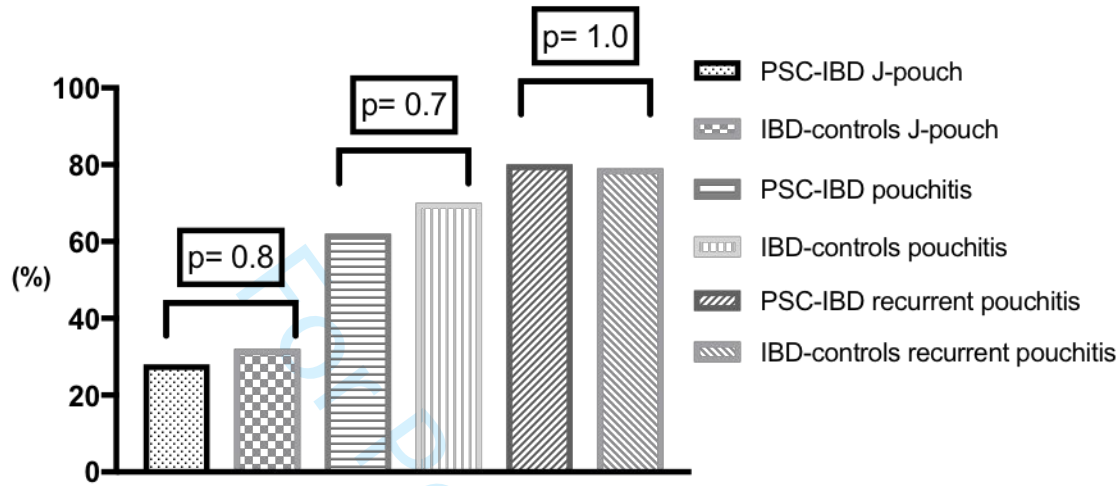
PSC: primary sclerosing cholangitis, IBD: inflammatory bowel disease

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